ISSN : (Online): 3008-038X
Abbreviation: J Derm Cse Rep
DOI: 10.61705/jdcr.19.2.2026.18.22

Journal of Dermatological Case Reports

Lymphangioma Circumscriptum of the Face: A Rare Entity

Bashaer Almahdi[1], Seham Marghalani[1], Reshale Johar[1], Hatoon Althobaiti[1], Talah Alturkistani[2], Lama Malibari[2]

[1] Dermatology Department, Ministry of the National Guard - Health Affairs, Jeddah, Saudi Arabia. King Abdullah International Medical Research Center, Jeddah, Saudi Arabia. King Saud bin Abdulaziz University for Health Sciences, Jeddah, Saudi Arabia.

[2] King Saud Bin Abdulaziz University for Health Sciences, Jeddah, Saudi Arabia.Email : talahqari@gmail.com

Corresponding Author

Bashaer Almahdi

Dermatology Department, Ministry of the National Guard - Health Affairs, Jeddah, Saudi Arabia. King Abdullah International Medical Research Center, Jeddah, Saudi Arabia. King Saud bin Abdulaziz University for Health Sciences, Jeddah, Saudi Arabia.

Keywords:

Lymphangioma circumscriptum; Microcystic lymphatic malformation; Facial lymphatic malformation; Cutaneous lymphatic malformation; Rare dermatologic condition; Case report

Abstract:

Lymphangioma circumscriptum, also known as microcystic lymphatic malformation, is a rare benign lymphatic anomaly that typically presents in early childhood and most commonly involves the trunk, axillae, and genital regions. Facial involvement is uncommon and may pose diagnostic challenges due to its atypical location and resemblance to other vesiculobullous conditions. A 14-year-old female presented with an 8-year history of asymptomatic vesicular lesions localized to the right zygomatic region. Examination revealed grouped translucent vesicles with a frogspawn appearance. Histopathologic evaluation showed dilated dermal lymphatic channels with lymphoid aggregates, and immunohistochemistry was positive for D2-40, confirming the diagnosis. This case highlights a rare facial presentation of lymphangioma circumscriptum and underscores the importance of considering lymphatic malformations in the differential diagnosis of chronic vesicular facial lesions. Histopathologic and immunohistochemical evaluation remains essential to ensure accurate diagnosis and appropriate management.

Received : 24-03-2026 **Revised : 02-04-2026** **Accepted: 02-03-2026** **Published : 06-04-2026**

Introduction

Lymphatic malformations (LM) are benign abnormalities of the lymphatic system that may be due to developmental anomalies or secondary to obstruction or damage of previously normal lymphatics. Whether congenital or acquired, these malformations result in lymphatic vessel dilation and fluid accumulation due to a disruption in the drainage process [1]. Lymphatic fluid accumulation gives rise to variable-sized cysts that may occur within the skin, mucous membranes, muscles, bone, or occasionally viscera. According to the size of the cysts, LMs are divided into microcystic, macrocystic, or mixed malformations, with the former being the most common [1]. Microcystic lymphatic malformation (MLM), also known as lymphangioma circumscriptum (LC), presents clinically as well-circumscribed clusters of translucent or hemorrhagic vesicles, typically

ISSN : (Online): 3008-038X
Abbreviation: J Derm Cse Rep
DOI: 10.61705/jdcr.19.2.2026.18.22

Journal of Dermatological Case Reports

appearing at birth or within the first few years of life. [2]. Lesions are commonly confined to the abdomen, axillae, mouth, and genital region. Cutaneous facial involvement in LC is rare; in published series, head and neck involvement is amongst the least commonly involved sites [3]. Associated symptoms of LC may include pruritus, pain, burning, lymphatic drainage, and infection. Lesions involving the face may also raise aesthetic concerns in patients and impact emotional well-being. Recognizing these rare anatomic presentations is pivotal for accurate diagnosis and appropriate management.

Case Presentation

A 14-year-old female presented with an 8-year history of fluid-filled lesions over the right side of the face. The lesions were relatively asymptomatic, with only occasional leakage of clear fluid upon manipulation. The patient denied any associated itching, bleeding, purulent discharge, ulceration, pain, rapid changes, or systemic symptoms. Although minimally symptomatic, the lesions were cosmetically distressing. According to the patient's parents, the lesions began as a few erythematous papules which gradually evolved to become vesicular in morphology. There was no preceding infection, inflammation, rash, trauma, surgery, radiation or new medication use. The patient briefly used topical tretinoin 0.025% and fusidic acid cream, with no clinical improvement. No systemic therapies were used for the lesions. Medical history was unremarkable. Family history was significant for atopy, as her mother had asthma; however, there was no history of a similar presentation or other dermatologic conditions. Upon dermatological examination, there were multiple grouped vesicular and papulovesicular lesions localized over the right zygomatic region. The lesions are translucent to pinkish in color, with some showing a slightly reddish or crusted appearance. They range in size from 2 to 5 mm, with smaller dome-shaped vesicles and a few larger coalescing clusters displaying the characteristic appearance of "frogspawn". The surrounding skin shows mild erythema without signs of acute inflammation or induration [Figure 1]. No secondary infection, ulceration, or significant scaling is noted. Differential diagnoses of atypical herpes simplex virus (HSV), bullous impetigo, and lymphangioma circumscriptum were considered. Skin biopsy confirmed a lymphatic malformation, showing several irregular thin-walled vascular channels in both the papillary and reticular dermis with evident lymphoid aggregates [Figure 2]. No acute inflammation or viral inclusions were identified. Immunohistochemical staining demonstrated positivity for D2-40 [Figure 3]. The patient was treated with pulsed dye laser therapy (fluence, 6.0 J/cm²; pulse width, 2.0 ms; pulse rate, 1.0 Hz; 9 pulses).

Discussion

Facial involvement in microcystic lymphatic malformation, also termed lymphangioma circumscriptum (LC), is rare with most reported cases occurring in the trunk genital, and perianal regions followed by the extremities and oral cavity [3]. In the largest clinicopathological series by Fatima et al., head and neck involvement constituted only a minority of cases, highlighting the unusual anatomical distribution observed in our patient [3]. Preach et al. described two variants of lymphangioma circumscriptum—classical and localized—with the classical variant characterized by an earlier age of onset and more extensive tissue involvement; lesions most frequently occur on the neck, axilla, breast, chest, and buttocks [4]. Most cases of LC present at birth or during early childhood, consistent with the onset of lesions at six years of age in our patient [4]. Several reports have described a slow, progressive course over many years with minimal symptoms, mirroring the indolent evolution observed in our case [5,6]. Clinically, LC typically presents as clusters of translucent or hemorrhagic vesicles with a characteristic "frogspawn" appearance, a hallmark feature consistently described in the literature [7,8]. Occasional leakage of clear lymphatic fluid has also been reported and was observed in our patient, further supporting the diagnosis [9]. However, atypical presentations have been documented, with clinical features overlapping those of other dermatologic conditions, including herpes zoster, molluscum contagiosum, viral warts, angiosarcoma, hemangioma, epidermal nevus, angiokeratoma, and leiomyoma [10,11]. A similar diagnostic challenge was encountered in our case, in which LC was included in the differential diagnosis and subsequently confirmed by histopathologic examination. Immunohistochemistry plays a crucial

ISSN : (Online): 3008-038X
Abbreviation: J Derm Cse Rep
DOI: 10.61705/jdcr.19.2.2026.18.22

Journal of Dermatological Case Reports

role in confirming the diagnosis, as the monoclonal antibody D2-40 is a highly sensitive and specific marker for lymphatic endothelial cells and is widely used to differentiate lymphatic malformations from other vascular lesions [12]. The histopathologic and immunohistochemical findings in our case, including D2-40 positivity, were consistent with previously reported cases and confirmed the diagnosis of lymphangioma circumscriptum [12]. Management of lymphangioma circumscriptum remains challenging, as surgical excision is considered the most definitive treatment when feasible; however, recurrence rates of up to 23% have been reported, particularly in cases with superficial or extensive involvement [13,14]. Various nonsurgical modalities—including laser therapies, electrosurgery, cryotherapy, and sclerotherapy—have been described with variable success, largely in case reports and small case series [13–15]. In our patient, pulsed dye laser therapy was initiated (fluence, 6.0 J/cm²; pulse width, 2.0 ms); however, only a single session was performed due to the unavailability of a functioning PDL system.

Conclusion

In conclusion, reporting such rare presentations expands current understanding of the disease spectrum and reinforces the importance of considering lymphatic malformation in the differential diagnosis of chronic vesicular facial lesions. Histopathologic examination remains essential for establishing the diagnosis, particularly in cases with atypical location, to prevent misdiagnosis and avoid unnecessary interventions.

References

1. Wassef M, Blei F, Adams D, Alomari A, Baselga E, Berenstein A, et al. Vascular anomalies classification: recommendations from the International Society for the Study of Vascular Anomalies. *Pediatrics*. 2015 Jul;136(1):e203–14. doi:10.1542/peds.2014-3673
2. Whimster IW. The pathology of lymphangioma circumscriptum. *Br J Dermatol*. 1976 May;94(5):473–86. doi:10.1111/j.1365-2133.1976.tb05134.x
3. Fatima S, Uddin N, Idrees R, Minhas K, Ahmad Z, Ahmad R, et al. Lymphangioma circumscriptum: clinicopathological spectrum of 29 cases. *J Coll Physicians Surg Pak*. 2015 Sep;25(9):658–61
4. Rao BA, Srikanth J, Muvva SH, et al. Lymphangioma circumscriptum – a rare case. *Int J Health Sci Res*. 2018;8(3):291–4
5. Bouabdella S, Dikhaye S, Zizi N. Lymphangioma circumscriptum of the scrotum: case report. *Ann Med Surg (Lond)*. 2021 Oct 15;71:102962. doi:10.1016/j.amsu.2021.102962
6. Sahu T, Mankar P, Soni R. Cutaneous lymphangioma circumscriptum. *J Med Excell*. 2024 Jul–Dec;1(1):23–4. doi:10.4103/JOME.JOME_4_24
7. Mehrotra K, Kumar JV, Kumar P. Giant lymphangioma circumscriptum: a differential of verrucous lesions. *Sch J Med Case Rep*. 2015;3(2):121–3. doi:10.36347/sjmcr.2015.v03i02.017
8. Bhattacharyya A, Sahadevan G. Frog-spawn appearance: diagnostic clue to lymphangioma circumscriptum. *CosmoDerma*. 2023;3:137
9. Gowda VRR, Arumugam VS, Rajaram MK, Govindharaju N, Tanaji ME. Lymphangioma circumscriptum on the buccal mucosa: a case report of a rare entity. *Cureus*. 2024 Sep 29;16(9):e70466. doi:10.7759/cureus.70466
10. Gomides MDA, Costa LD, Berbert ALCV, Janones RS. Cutaneous lymphangioma circumscriptum: the relevance of clinical, dermoscopic, radiological, and histological assessments. *Clin Case Rep*. 2019 Apr;7(4):612–5. doi:10.1002/ccr3.2007
11. Bidaki R. A case of long-lasting misdiagnosed lymphangioma circumscriptum masquerading as dermatitis. *Int J Dermatol*. 2018
12. Sadasivan A, Ramesh R. Lymphangioma circumscriptum – a rare cause of gingival enlargement: a case report and review of literature. *Clin Cosmet Investig Dent*. 2022;14:199–206. doi:10.2147/CCIDE.S367281
13. Vignes S, Arrault M, Trévidic P. Surgical resection of vulva lymphoedema circumscriptum. *J Plast Reconstr Aesthet Surg*. 2010 Nov;63(11):1883–5. doi:10.1016/j.bjps.2009.11.019
14. Farnoosh S, Don D, Koempel J, Panossian A, Anselmo D, Stanley P. Efficacy of doxycycline

ISSN : (Online): 3008-038X
Abbreviation: J Derm Cse Rep
DOI: 10.61705/jdcr.19.2.2026.18.22

Journal of Dermatological Case Reports

and sodium tetradecyl sulfate sclerotherapy in pediatric head and neck lymphatic malformations. *Int J Pediatr Otorhinolaryngol*. 2015 Jun;79(6):883–7. doi:10.1016/j.ijporl.2015.03.024

15. Emer J, Gropper J, Gallitano S, Levitt J. A case of lymphangioma circumscriptum successfully treated with electrodessication following failure of pulsed dye laser. *Dermatol Online J*. 2013 Mar 15;19(3):2

Figure Legends

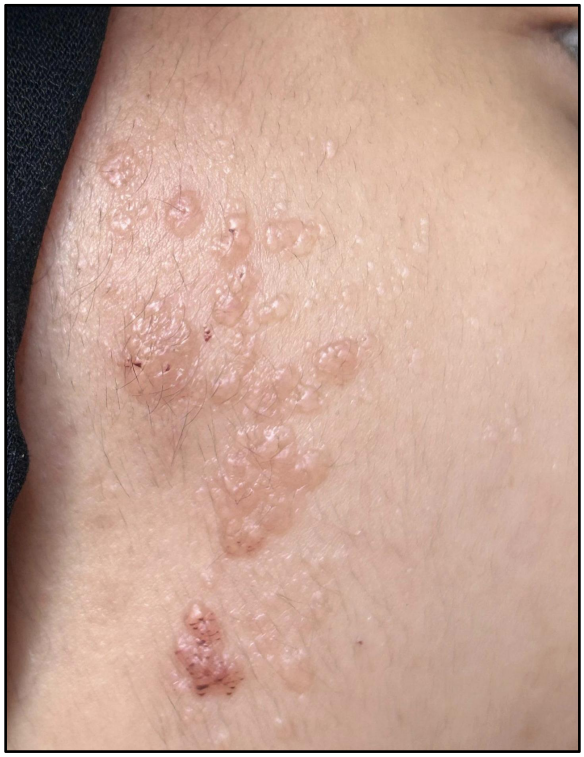

Figure 1. Grouped translucent to pinkish vesicles and papulo-vesicles (2–5 mm) over the right zygomatic region, some coalescing into a characteristic ‘frogspawn’ pattern with mild erythema.

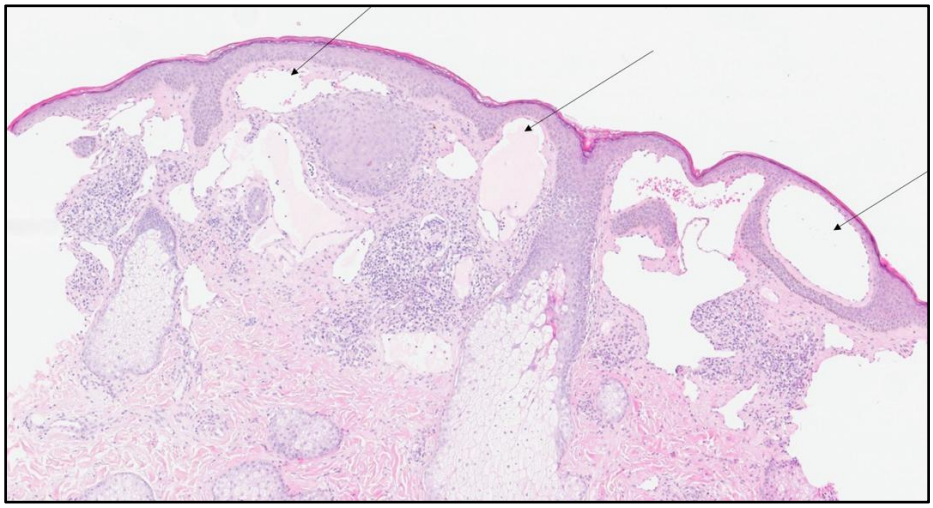

Figure 2. Hematoxylin and eosin–stained section showing multiple dilated, thin-walled lymphatic channels within papillary and reticular dermis (arrows), consistent with lymphangioma circumscriptum.

ISSN : (Online): 3008-038X
Abbreviation: J Derm Cse Rep
DOI: 10.61705/jdcr.19.2.2026.18.22

Journal of Dermatological Case Reports

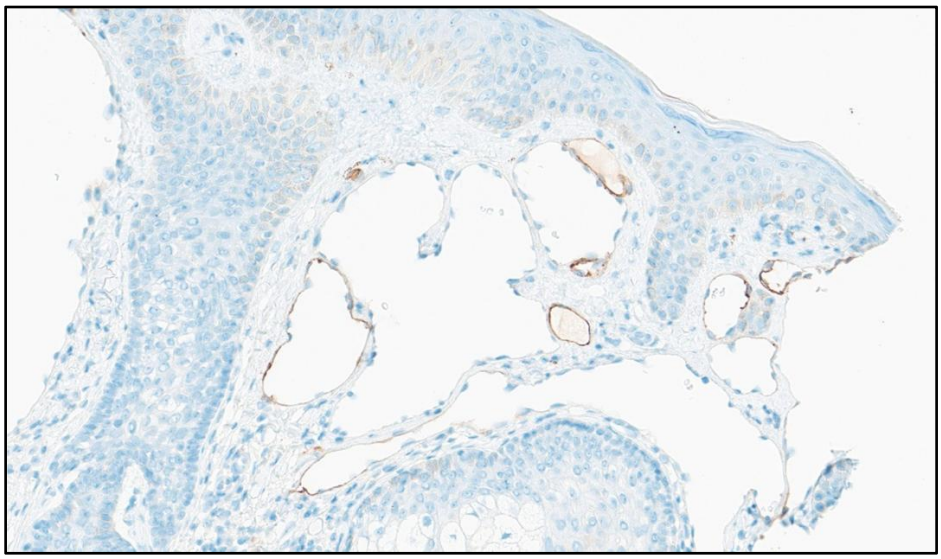

Figure 3. Immunohistochemical staining with D2-40 highlighting lymphatic endothelial cells lining the dilated dermal channels, confirming their lymphatic origin (brown staining).